"CASE SERIES OF CARDIAC MYXOMAS – A SINGLE INSTITUTIONAL EXPERIENCE"

Dissertation submitted for

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BRANCH I – CARDIOTHORACIC SURGERY

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"learn to heal"

CERTIFICATE

This is to certify that the dissertation entitled **"CASE SERIES OF CARDIAC MYXOMAS – A SINGLE INSTITUTIONAL EXPERIENCE"** is the bonafide original work of **DR.M. RAJAN** in partial fulfillment of the requirements for M.Ch Branch-I CARDIOTHORACIC SURGERY examination of THE TAMILNADU DR.M.G.R. MEDICAL UNIVERSITY to be held in August 2008.The period of post-graduate study and training was from August 2005 to July 2008.

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DECLARATION

I **Dr.M. RAJAN,** solemnly declare that this dissertation entitled, **"CASE SERIES OF CARDIAC MYXOMAS – A SINGLE INSTITUTIONAL EXPERIENCE"** is a bonafide work done by me at the Department of Cardiothoracic Surgery, Madras Medical College and Government General Hospital during the period 2005 – 2008 under the guidance and supervision of the Professor and Head of the Department of Cardiothoracic Surgery, Madras Medical College and Government General Hospital, Professor **K. Harshavardhan M.S., M.Ch.** This dissertation is submitted to The Tamil Nadu Dr.M.G.R Medical University, towards partial fulfillment of requirement for the award of **M.Ch. Degree (Branch – I) in Cardiothoracic Surgery**

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Dr.M. RAJAN

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CONTENTS

Introduction

Cardiac tumours include benign and malignant neoplasms arising within the cardiac chambers or in the myocardium.Approximately 70% of cardiac tumours are benign and 30% are malignant and potentially capable of invasion or metastasis[1].

Cardiac myxomas are primary cardiac tumours that are generally pedunculated but may have a broad base .They are intracavitary tumours occurring within any of the cardiac chambers, but they have a predilection for the atria, particularly the left. They are usually 5-6 cm in diameter, with a range of 1-15 cm.

Most atrial myxomas, whether left or right, arise from the atrial septum , usually from the region of limbus of the fossa ovalis. About 10% have other sites of origin particularly posterior and anterior atrial walls and the appendage. Most myxomas 80%-90% are in the left atrium. Right atrial myxomas tend to be more solid and sessile than left atrial myxomas, with a wider attachment to the atrial wall or septum.[2] Myxomas may

occasionally be found in right ventricle on the free wall or ventricular septum. Few cases of left ventricular myxomas are also reported . Valvar myxomas arising from mitral, tricuspid and pulmonary valves have also been reported.

Myxomas may produce symptoms of hemodynamic derangement from obstruction of flow within the cardiac chamber, symptoms associated with embolization and constitutional symptoms.

HEMODYNAMIC DERANGEMENT

Myxomas may obstruct pulmonary or systemic venous drainage or may impair flow across the atrioventricular valves .The obstruction is characteristically progressive. When obstruction is intermittent , syncope, often related to postural change, or sudden death may occur. Impairment of valve closure ,either by obstruction or leaflet damage ,may cause regurgitation. Although regurgitation is the dominant abnormality in a few patients, as a rule obstruction predominates.

EMBOLISM

A major feature of cardiac myxomas is embolization. Emboli may arise from tumour fragmentation or detachment of entire tumour, or from thrombi or infected foci on the neoplasm. Systemic emboli occur in 30%-45% of patients with left atrial myxomas .Embolism from right sided tumours occurs in about 10% of cases and may cause massive fatal pulmonary obstruction. Massive pulmonary embolism complicating left atrial myxoma has been reported [3].

CONSTITUTIONAL SYMPTOMS

In about 30% of patients , the only manifestation of cardiac myxoma are a plethora of constitutional symptoms. Large left atrial myxomas are particularly apt to produce constitutional symptoms. These symptoms include fever, weight loss, clubbing, raynauds phenomenon, myalgia and arthralgia. Other unusual manifestations include polycythemia, hemolytic anemia.

Nearly all solitary myxomas are non familial, myxomas have familial occurrence in about 5% of patients. Familial myxomas have autosomal dominant inheritance and are primarily disorders

of young men. Familial myxomas are associated with serotoli cell tumour of testis, cushings syndrome, pituitary tumours, centrofascial and labial lentiginosis, cutaneous myxomas and multiple myxoid mammary fibroadenomas. Familial myxomas have the same histologic appearance as nonfamilial myxomas and produce the same symptoms, however they have a strong tendency to recur.

Investigations to diagnose cardiac myxomas are largely dependent on Transthoracic echo (TTE) supplemented by Transesophageal echocardiography (TEE) .Cardiac catheterization and angiography no longer constitute the investigation of choice unless other types of cardiac or coronary artery disease require assessment.

Although rhabdomyoma is by far the most common primary heart tumour in children and infancy, cardiac myxomas are the most commonly encountered primary heart tumour in adults. In this retrospective study we analyse the incidence, presentation and operative management of cardiac myxomas in the Department of Cardiovascular and Thoracic surgery, Madras Medical College, Chennai between the years 2003 and 2008.

Review of Literature

DEFINITION

Cardiac myxomas are primary cardiac tumours that are generally pedenculated but may have a broad base. Cells are uniform, small and polygonal with round or oval nuclei and a moderate amount of cytoplasm. They lie in a myxomatous stroma in which other elements may be seen. One feature that distinguish them from thrombi is that they are covered by endothelium- lined crevices and cleft4

INCIDENCE OF CARDIAC MYXOMAS

Cardiac myxomas comprise 29% of neoplasms of heart and pericardium [5]. Most myxomas 90% are in the left atrium. Right atrial myxomas tend to be more solid and sessile than left atrial myxomas with a wider attachment to the atrial wall or septum[6]. Ventricular myxomas are rare and are found to arise mostly from right ventricular freewall or ventricular septum. In 15% of reported cases ,right ventricular myxomas are associated

with other cardiac myxomas .Left ventricular myxomas are rare , and little information is available about them[7] Valvar myxomas arising from mitral, tricuspid and pulmonary valves have been reported.[8,9,10].

Mcalister and Fenoglio et al[1] in their published series of tumours of cardiovascular system have found cardiac myxomas comprised 29%, lipomas 10%, papillary fibroelastomas 9.5% and rhabdomyoma 8% of all cardiac neolasms. In the malignant forms angiomyosarcoma predominated with 8.8%, rhabdomyosarcoma with 5.8%.

Sutsch et al [14] in their published series of occurrence, type and location of cardiac tumours as detected by Doppler echocardiography are reviewed. In their series of 20,305 consecutive echocardiographic studies performed at our institution over a four year period, cardiac tumours were identified in 30 patients (0.15%). Primary cardiac tumours were detected in 21/30 (70%) , secondary tumours in 5/30 (17%) , however in 4/30 patients (13%) the tumours could not be classified. In the primary tumour group, benign cardiac tumours were found in 18/21 patients

(86%) and malignant tumours in 3/21 (14%). All benign cardiac masses were myxomas (18/18) and accounted for 60% of all tumours. They were mainly located in the left atrium 16/18 (89%) and rarely occurred in the right atrium 2/18 (11%).

CELL OF ORIGIN

Myxomas are composed of cells, primitive capillaries and foci of extramedullary hematopoiesis within a myxoid matrix of acid mucopolysaccharide. The stroma contains variable numbers of reticulocytes and elastin fibres, smooth muscle cells and collagen deposits. The matrix also contains polygonal cells with scant eosinophilic cytoplasm ,either single and stellate or multinuclear and in small nests. At the periphery of the tumour the cells form a monolayer with clustering in the crevices, thereby simulating primitive capillaries .The stalk has abundant large arteries and veins that communicate with the subendocardium, and at this interface, lymphocytes and plasma cells are prominent as studied by Merkow et al [11].

Hurst JW et al[12] found that the notion that myxomas are derived from thrombi has been thoroughly dispelled .He found

that the nucleus of the polygonal cells are typically irregular and slightly hyperchromatic, but mitosis are not seen. The cells contain fine parallel filaments similar to those seen in glioma and fibromyxosarcoma. These filaments are believed to be the contractile components of smooth muscle cells. Immunologically identifiable smooth muscle-type filaments are recognized in endocardial cells, which are more abundant in the left atrium, especially in the region of fossa ovalis, than in other chambers.

Ferrans and Roberts et al[13] in their study made that the organelle content of myxoma cells does not provide sufficient information to determine the cell of origin .Although myxoma cells have a vasoformative tendency , in their view the cytoarchitectural features of the variously differentiated blood – vessel like structures differ from those of normal blood vessels. Thus myxomas tend to arise from multipotential mesenchymal cells capable of differentiating into various types of cells, a view supported by the finding of bone and bone marrow tissue in myxomas. Histologic examination of the atrial septum in 11 autopsied patients younger than age 4 months revealed myxomatous or myxofibrous tissues in the endocardium near the

fossa ovalis, further supporting the concept that myxomas are derived from embryonal , undifferentiated mesnchyme.

Vaideeswar and Butany et al[15] in their study found that cardiac myxomas arise from pluripotent mesenchymal cells and are seen as intracardiac, glistening polypoid masses arising from most frequently from the interatrial septum(IAS) in the left atrium. They are composed of stellate to polygonal myxoma cells in a mucopolysaccharide-rich matrix. These tumors can be sporadic or familial.

Basson et al[16] in his study after extensive analysis ultimately confirmed that these lesions represented a primary neoplastic process. Although the cell of origin has yet to be isolated, they have been posited to arise from a subendocardial cell, termed “reserve” or “lepidic” cell. These tumors usually exhibit a typical hypocellular appearance of small pyramidal or stellate cells against a bland proteoglycan background. They may also exhibit evidence of a wide variety of cellular lineages, including zones of extramedullary hematopoiesis, acinar structures suggestive of epithelial organization, cells with electron

microscopic features suggestive of muscle, and a number of immunohistochemical markers consistent with a remarkable range of cell types. Such histological studies suggest that the cardiac reserve cell is pluripotent and, in the appropriate genetic and biochemical milieu, may be a progenitor cell for a number of cardiac cells in the healthy and pathological heart.

Tetsuya Kono et al[17] in their study found the association between angiogenesis and the clinicopathologic features in cardiac myxoma, vascular endothelial growth factor (VEGF) in the myxoma was examined by using reverse transcriptase polymerase chain reaction and immunohistochemistry, and the micro vessel density was determined by counting micro vessels in the myxoma by using immuno staining for platelet endothelial cell adhesion molecule (P-ECAM). They found that there was an inverse correlation between the tumor size and the ratio of the micro vessel density in the central part to the micro vessel density in the peripheral part of myxoma. Furthermore, there was an inverse correlation between the proliferating cell nuclear antigen-labeling index and the tumor size, and the proliferating cell nuclear antigen-labeling index in myxomas with high

vascular endothelial growth factors expression was higher than that in myxomas with low VGEF expressions.

SYMPTOMS IN CARDIAC MYXOMA

There are three major syndromes linked to myxomas: Embolic events, Obstruction of blood flow and Constitutional symptoms. Embolic events happen when fragments of the tumour, or the thrombi attached to the outside of the tumour, are released and enter the blood stream .Gelatinous myxomas are more likely to embolize than the solid form of this tumour.

Myxomas also may obstruct blood flow in the heart, usually at a heart valve. The mitral valve is the heart valve most commonly affected. Blood flow restrictions can lead to pulmonary congestion and heart valve disease .Embolization can lead to severe consequences. In cases of left atrial myxomas 40-50% of patients experience embolization. Emboli usually end up in brain kidneys and extremities.

The third syndrome linked to myxomas are called constitutional syndromes, non specific symptoms caused by the myxoma.

Scott and Veinot et al[18] reported their analysis of clinical, pathologic, and echocardiographic features of cardiac myxomas that were surgically removed between 1976 and 1999 at the university of Ottawa Heart Institute. There were 34 patients (mean age, 53 years). Of these 25 patients had symptoms, and the common symptoms were dyspnoea in 13 patients and embolism in 9 patients. Nine patients were asymptomatic.

Piazza et al[19] reported their experience at McGill university teaching hospitals, Montreal, Quebec. 21 patients resected for primary cardiac tumors were reviewed retrospectively in terms of clinical presentation, surgical treatment, histopathological findings and outcome. The most common clinical presentation in adults was dyspnoea (38%), cenral nervous system /embolic phenomena (24%), and in the pediatric age group, it was hypoxia (50%).

Keeling, Oberwalder et al[20] in their experience at Karl Franzens University Austria operated on 49 consecutive patients

with cardiac myxoma. In their series most myxomas originated from the left atrium, 6.1% from the mitral valve, 4.1% from the right atrium and 2% had biatrial myxomas. Cardiac signs appeared in 93.9% of the patients and embolic events occurred in 26.5%.

Percell et all[21] in their study at Department of Medicine James A Haley VA Hospital, Tampa, FL found that atrial myxomas are the most common primary tumor of the heart and occur in as many as 3 in 1000 pateints. These tumors are a major cause of patient morbidity and mortality. Approximately 50% of patients with myxomas may experience symptoms due to central or peripheral embolism or intracardiac obstruction, but 10% of patients were completely asymptomatic. They were diagnosed to have cardiac myxomas when they were investigated routinely.

MacGregor and Cullen et al[22] in 1959 were the first to report the systemic manifestations of fever , weight loss, anemia, increased erythrocyte sedimentation rate and elevated globulins in patients with cardiac myxoma. These findings are completely

reversible after removal of the tumor 23. The elevated gamma globulins suggests antibody formation perhaps due to myxomatous emboli, but what causes these systemic symptoms is unknown.

Koo Cho et al[24] in their experience of operating 20 patients of cardiac myxomas at Severance Hospital , Seoul ,Korea 95% (19/20) of the patients were symptomatic .They had either dyspnea on exertion ,palpitation, paroxysmal nocturnal dyspnea, chest pain and neurological symptoms. One patient was asymptomatic.

INVESTIGATIONS FOR DIAGNOSIS

IMAGING STUDIES

Chest Radiography

- Abnormal cardiac silhouette , mimicking mitral stenosis
- Unusual intracardiac tumour calcification
- Pulmonary edema

ECHOCARDIOGRAPHY

Although TEE is more sensitive , 2-dimensional echocardiography is usually adequate for diagnosis.

Tumour location, size, attachment and mobility can be assessed with this technique.

An atrial myxoma must be differentiated from a left atrial thrombus.The thrombus is usually situated in the posterior portion of the atrium and has a layered appearance. Presence of a stalk and mobility favours atrial myxoma.

Doppler echocardiography can show the hemodynamic consequences of atrial myxoma.

TRANS ESOPHAGEAL ECHOCARDIOGRAPHY

- Better specificity and 100% sensitivity compared to transthoracic echocardiography.
- Good resolution of both atria and atrial septum.
- Better visualization of anatomic details of the tumour and stalk.
- Reveals smaller (1-3mm) diameter tumours.
- Visualizes atrial appendages.
- Detects shunting
- Advisable for myxoma syndrome-multiple less common sites.

MAGNETIC RESONANCE IMAGING

MRI provides useful information about size, shape and surface charecterstics on T1- weighted images. Cine MR gradient echo (GRE) images can demonstrate mobility of the tumour. Point of attachment is best visualized by MRI with a [ost surgical correlation of 83%. In a small series , MRI was superior to CT scan ,which showed only 30% correlation for the site of attachment.

Information about tissue composition can be used to differentiate a tumour and a thrombus.

CARDIAC CATHETERIZATION

- Used only in selected patients in whom non invasive evaluation is inadequate.

- Performed to exclude coexistent coronary artery disease in patients more than 40.

- Was the pre- echocardiographic method of diagnosis.

Anderson et al[25] and Smellie et al[26] in their study found that those patients with obstructing type of left atrial myxoma , their chest roentgenograms closely resembles mitral valve disease. Kerley B lines have been reported, as has calcification of the mitral valve. The electrocardiogram is non-specific in left atrial myxoma and only reflects the anatomic changes secondary to the tumour. Atrial fibrillation is present in only 14% of patients 27.

Koo Choo et al[24] in their surgical experience of 20 cases of atrial myxomas at The Severance Hospital , Seoul their first 4 patients were tested with angiocardiography alone, and 3 of these were misdiagnosed. The last 16 were tested by

angiocardiography, M- mode echocardiography, and 2-dimensional echocardiography, alone or in various combinations, and there were no further misdiagnoses. From their experience they concluded that 2-dimensional echocardiography was the most accurate method of diagnosing cardiac tumours.

Piazza et al[19] in their retrospective study on 21 patients resected for primary cardiac tumours, all but one of the tumours were visualized using transthoracic echocardiography. One patient required angiography for its diagnosis. They concluded from their study that 2-dimensional echocardiography was accurate in diagnosing cardiac myxomas.

Acebo et al[28] in their experience of 34 patients 25 patients were symptomatic, 9 were asymptomatic. All patients were diagnosed by 2-dimensional echocardiography which corroborated well intraoperatively .They concluded that apart from reliability in diagnosis it provides additional insight regarding the potential for embolism.

Agarwal et al[29] reported their experience with 34 patients with primary cardiac tumours. Of these 31 patients were found to

have myxoma , left ventricular fibroma in 1, and leiomyosarcoma in 2.Echocardiography was diagnostic in all the benign tumours, whereas the malignant tumours were found incidentally during surgery.

Percell et al[21] from their experience at James A Haley VA Hospital reported that screening for myxomas should involve a thorough history, physical examination and a transthoracic and/or transesophageal echocardiogram .Transthoracic echocardiogram was 95% sensitive in detecting cardiac myxomas, and transesophageal echocardiography approaches 100% sensitivity.

SURGICAL APPROACHES FOR CARDIAC MYXOMAS

Once the diagnosis of cardiac myxoma has been made, surgery should be done as soon as possible. An 8% mortality has been reported in patients awaiting operation following definitive diagnosis 30. Cardio-pulmonary bypass must be established promptly, a coexisting valvular lesion should be looked for and all cardiac chambers and all main adjacent cardiac vessels must be visualized to detect synchronous tumors. Complete cure may be expected, especially if the involved atrial septum is excised and repaired by patch replacement to avoid tumor recurrence31.

Piazza et al[19] in their series of 21 cases surgical approach was uniatrial in 10 patients (48%)-seven (33%) via the left atrium and three (14%) via right atrium. Eight patients (38%) required biatrial approach; the remaining three patients (14%) with ventricular tumours required ventricular approach. Three valve myxomas ,two involving the mitral valve and one involving tricuspid valve, were encountered; all underwent conservative leaflet resection and valve repair.

Koo Choo et al[24] in their experience of 20 cases (19 cases of LA myxomas, 1 case of RA myxoma), all the LA myxomas were approached through the left atrium via a median sternotomy and cardio-pulmonary bypass. Before resection, all other cardiac chambers were explored for multicentric tumours. The pedicle was then excised completely along with a cuff of tissue to which it was attached. The surgically created atrial septal defect was closed by direct suture alone in 10 instances, by prosthetic patch in 8 instances and in 2 instance by direct suture and patch placement.

Baksaas et al[32] in their series of 30 patients with cardiac tumours 27 patients had benign tumours, 25 of which were myxomas predominantly located in the left atrium. All myxomas were operated in right atrium and the inter atrial septum was excised with the myxoma mass. All the other chamber were examined for synchronous tumour and the inter atrial septum was closed with patch.

May et al[33] in their series used biatrial approach for all cases of left atrial myxoma. The usual incision was made in the

left atrium posterior to the interatrial groove. The point of attachment of the tumour to the atrium was determined by inspection and exploration of the tumour with the index finger. An oblique right atriotomy was made and the interior of the right atrium examined in case a second tumour is present. The IAS was then opened with a knife near the center of the fossa ovalis, and with the finger in the left atrium as a guide to the attachment of the tumour, a sufficient amount of atrial septum is excised to include the tumour attachment and if possible uninvolved tissue 5 mm beyond it. The defect in the IAS was closed by direct technique or with a pericardial or PTFE patch.

The classical Dubost's technique of Bi-atrial approach is used in cases of large left atrial myxomas where a transverse right atriotomy is done which is extended onto the inter-atrial septum. Kabbani et al 41 have reported 24 cases of atrial myxomas approached through classical bi-atrial approach.

CONCOMITANT VALVAR INTERVENTIONS

The valves should be very thoroughly examined after removal of the tumour.Concomitant mitral valve procedures may range from mitral annuloplasty to valve repairs to valve replacements. A thorough pre-operative diagnosis and intra-operative assessment is needed. Kabbani et al [41] and Semb et al [42] have reported valve replacements in 4 cases out of their 24 reported cases.

IMMUNOLOGIC ABNORMALITIES IN CARDIAC MYXOMA

The histologic diagnosis is straight forward ,but the subject of discussion concerns the risk of recurrence after surgery.This risk has now been clearly identified and is correlated with young age, a family history of myxoma and multifocal lesions. More recently IL-6 and endothelial growth factor have been identified as markers for these tumours.

Mendoza et al [37 38] in their study have elaborated the role of interleukin-6 in the diagnosis, predicting the size and their role in detecting recurrences. Soeparwata et al [39] in their study reported similar correlation between tumour size and interleukin-6 plasma levels. Selkane et al [40] in their series of 40 cases with long term follow up have confirmed the role of interleukin-6 for detecting recurrences.

Aims and Objectives

Retrospective study to review and analyse the experience in diagnosis and surgical management with emphasis on the evolution at our institution of surgical techniques and treatment of cardiac myxomas.

Materials and Methods

30 consecutive cardiac surgical patients diagnosed with cardiac myxoma admitted in the Department of Cardiovascular and Thoracic Surgery , Madras Medical College, Chennai between April 1,2003 to March 30,2008 comprised the sample for this study .Patients operated before August 31,2005 were analysed from the case sheets obtained from Medical Record Department. A detailed clinical examination and findings were recorded over structured proforma (Annexure) for all patients .

All the patients under the study are classified according to their age, sex , mode of presentation , method of diagnosis, site of the tumour and surgical approach .

Observation and Results

OBSERVATION AND RESULTS

30 consecutive patients diagnosed with cardiac myxoma who underwent surgical management at The Department Of Cadiovascular and Thoracic Surgery, Madras Medical College, Chennai between April 1,2003 and March 31,2008 were retrospectively analysed. The subjects chosen in the study were classified into different groups based on their age and sex as follows (Table 1 and 2).

TABLE 1

S.NO	AGE	TOTAL
1	10-19	2
2	20-29	7
3	30-39	8
4	40-49	10
5	50-59	2
6	60-69	1
TOTAL		30

The highest number of observations were in the 40-49 years age group constituting more than 30% of the study group , followed by age group 30-39 years (8) constituting more than one fourth of the observations. 83% of the tumours were found to occur between 20-49 years age group.

TABLE- 2

AGE	MALE	FEMALE	TOTAL
10-19	0	2	2
20-29	2	5	7
30-39	4	4	8
40-49	2	8	10
50-59	0	2	2
60-69	0	1	1
TOTAL	8	22	30

Table 2 shows that 73% (22/30) of cardiac myxoma occurred in females and 27% (8/30) of the disease were found to occur in males and amongst the two sex groups , 77% and 100% of the disease occurred between 20-49 years in females and males respectively

TABLE-3

SYMPTOMS UPON PRESENTATION

CARDIAC SYMP	NEUROLOGIC SYMP	CONSTITUTIONAL SYM
30	2	8

Table 3 shows that all subjects (100%) included in the study presented with cardiac symptoms, only 7% (2/30) of the patients had neurological symptoms (one patient had left hemiparesis and the other had Freidrich's ataxia), 27% (8/30) of the patients had constitutional symptoms in the form of fever, anemia and raised erythrocyte sedimentation rate.

TABLE-4

MODE OF DIAGNOSIS

TTE	TEE	CAG
25	5	2

Table 4 shows that Trans Thoracic Echocardiography was diagnostic in 25 cases (83%) ,Trans Esophageal Echocardiography was required in diagnosing myxoma in 5 cases (17%). 2 cases underwent coronary angiogram for assessing the status of coronary arteries preoperatively and were found to have normal coronaries.

TABLE -5

SITE OF MYXOMA

LEFT ATRIAL	21
RIGHT ATRIAL	4
RIGHT VENTRICULAR	2
LEFT ATRIAL AND MV	2
MITRAL VALVE	1
TOTAL	30

Table 5 shows the location of cadiac myxomas in the cases included in the study. Myxomas were found in the left atrium in 21/30 cases (70%), Right atrial myxomas were found in 13% of cases.2 cases (7%) had Right ventricular myxomas. One case was found to have isolated mitral valve myxoma involving the posterior mitral leaflet and another two cases involving the left atrium and mitral valve.

TABLE-6

SITE OF PEDICLE ATTACHMENT TO ATRIAL WALL

LEFT ATRIAL SEPTUM INV. F.O	18
LEFT ATRIAL SEPTUM NOT INV F.O	05
FOSSA OVALIS IN RT.ATRIUM	04
MITRAL VALVE PML	01
TREBACULAR SEPTUM IN RV	01
RIGHT VENTRICULAR WALL	01
TOTAL	30

The sites to which the stalks of the atrial myxomas were attached are summarized in Table 6. Most commonly the attachment was to the fossa ovalis ,and with one exception (attachment to the Poterior mitral leaflet), the other neoplasms also involved the interatrial septum. Diameters of the atrial myxomas measured laterally and discounting pedicles, ranged from 2cm to 8cm. There were two right ventricular myxomas each involving the trebacular septum and right ventricular free wall.

TABLE -7

SURGICAL APPROACH AND TECHNIQUES

RIGHT ATRIAL	25
BI-ATRIAL	03
LEFT ATRIAL	01
RIGHT VENTRICULAR	01
TOTAL	30

Table 7 shows the surgical approach used for myxoma excision. Majority of the cases (83%) were removed by right atrial approach (25/30) cases. Bi-atrial technique was used for tumour excision in 3 cases, Left atriotomy was done in 1 case (removal of tumour arising from PML of MV). Right ventriculotomy was done for tumour retrieval in a case of RV myxoma.

TABLE -8

MEAN ACC TIME AND CPB TIME

S.NO	APPROACH	ACC TIME(MEAN)	CPB TIME(MEAN)
1	RT.ATRIAL	49.96’	86.76’
2	BI-ATRIAL	100.6’	153.0’
3	LEFT ATRIAL	108.0’	148.0’
4	RIGHT VENTR	66’	124’

Table 8 shows the mean Aortic cross clamp time and Total cardiopulmonary bypass time recorded for each approach .Short cross clamp time and cardiopulmonary bypass time were noted in right atrial approach.

In all 30 cases ,the atrial myxomas were excised through a median sternotomy, cardiopulmonary bypass. In all cases , cardiac arrest was achieved with antegrade cold crystalloid cardioplegia with topical hypothermia. Typically a right atrial incision was

made paralle l to right atrioventricular groove in 25 cases. The superior portion of the inter-atrial septum was incised to visualize the left atrial mass .Once the tumour was visualized from the right atrial aspect, the rest of the atrial septum was incised carefully avoiding cutting through the left atrial mass .Before resecting the other chambers were explored for the occasional multicentric tumour. The pedicle was completely excised along with a cuff of tissue to which it was attached. To minimize the risk of embolization , great care was taken to handle the pedicle gently ,and to remove any residual tumour debris from the surgical field.

The surgically created septal defect was closed by the following methods: in 22 instances , by pericardial patch closure with running monofilament suture. In 3 instances by direct closure.

Two patients required mitral valve replacement in addition to myxoma excision .One patient required mitral valve repair in addition to myxoma excision.

Discussion

PREVALENCE

Primary cardiac tumours are rare ,and 80% of them are benign. They account for 5% to 10% of all cardiac neoplasms, the balance of which are secondary tumours (therefore metastatic and malignant).Myxomas account for approximately 50% of all primary cardiac neoplasms ,and have been reported in all cardiac chambers. Although about 75% of myxomas occur in the left atrium and 25% in the right 34, multiple myxomas have been reported in 5% of patients with left atrial or left ventricular myxomas. Myxomas predominantly occur in women in the 3rd to 6th decades of life, and familial tendency have been reported 35.In

our experience myxomas were found to occur predominantly between 20-49 years age group (83%) as shown in Table -1. The tumour was found to occur predominantly in females in 73% and males in 27%, however in both sex groups the tumour occurred mostly between 20 -49 years as shown in Table-2.

CLINICAL MANIFESTATIONS

Although small tumours may be asymptomatic ,the clinical manifestations of atrial myxomas ,when they occur are well recognized. The most common findings are symptoms of valvular obstruction and signs of peripheral embolization. In our study group all patients 30/30 (100%) presented with any one of the cardiac symptoms in the form of dyspnea on exertion, palpitation, paroxysmal nocturnal dyspnea, chest pain and discomfort. This is

attributed to the fact that most cases seek medical attention only when they become symptomatic and screening for such cases are far from their reach. We encountered 1 case of atrial myxoma with systemic embolization and one case was associated with Freidrich's Ataxia. Constitutional symptoms such as fever, weight loss, arthralgia and anemia were noted in 27% of the cases (8/30) in our study as shown in (Table-3).

Although the exact incidence of chamber enlargement in chest roentgenography in patients with cardiac myxoma has not been described, chamber enlargement/hypertrophy was found to occur in 47% (14/30) of cases in our study.

DIAGNOSIS

Historically cardiac tumours have been diagnosed by angiocardiography and M-mode echocardiography. Transseptal angiocardiography has been used in in numerous centers, but tumour embolism has been reported with use of this method 36. Pulmonary levophase angiocardiography has proved a reasonably accurate method of visualizing a left atrial myxoma. Yet in our series, cardiac catheterization was not used as the sole radiological method of diagnosing a myxoma ,however 2 cases underwent catheterization for assessing their coronary artery status preoperatively.

In 25 of our patients, the cardiac mass was diagnosed preoperatively by Trans Thoracic Echocardiography. 5 patients with left atrial myxoma required Trans Esophageal Echocardiography

for the confirmation .In our study 2-dimensional echocardiography was diagnostic in 83% of cases and TEE was needed for confirmation in rest of the cases Surgical findings corroborated with the echocardiographic findings in all but one case where mxyoma was found to be adherent extensively to roof of left atrium, mitral valve and inter-atrial septum as shown in Table-6.

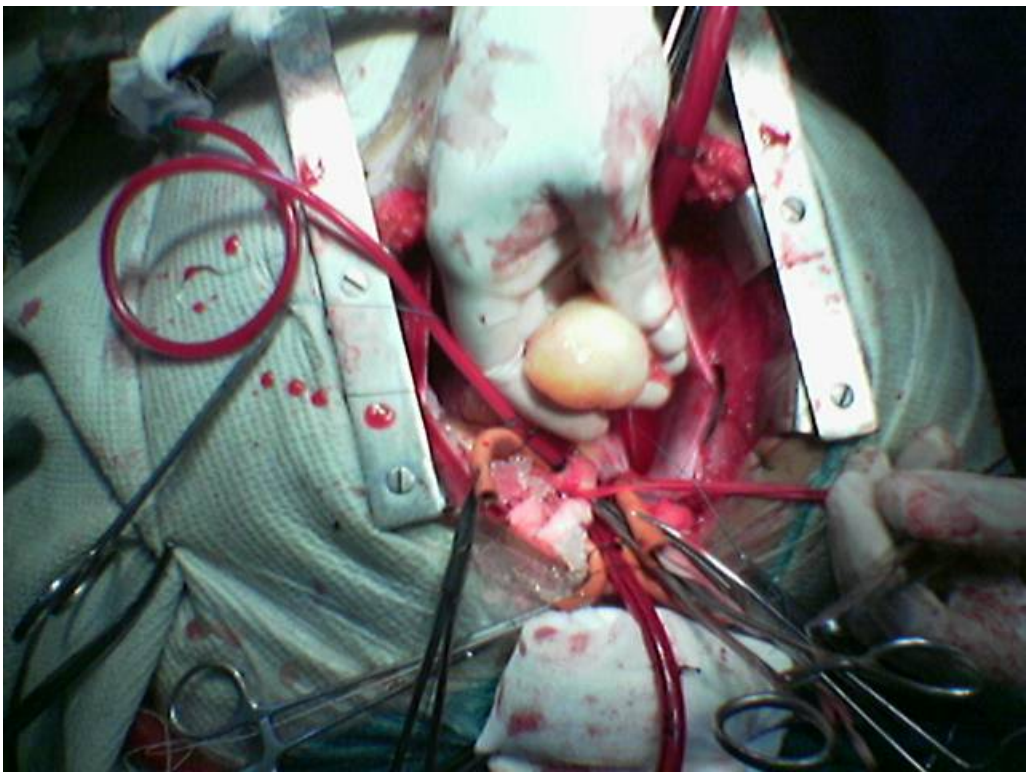
SURGICAL OBSERVATIONS AND RECOMMENDATIONS

When a diagnosis of left atrial myxoma is made, surgery should be done without delay .Current surgical techniques for treating atrial myxomas include median sternotomy with total cardiopulmonary bypass ,using moderate hypothermia with cold crystalloid cardioplegia with minimal touch technique of the

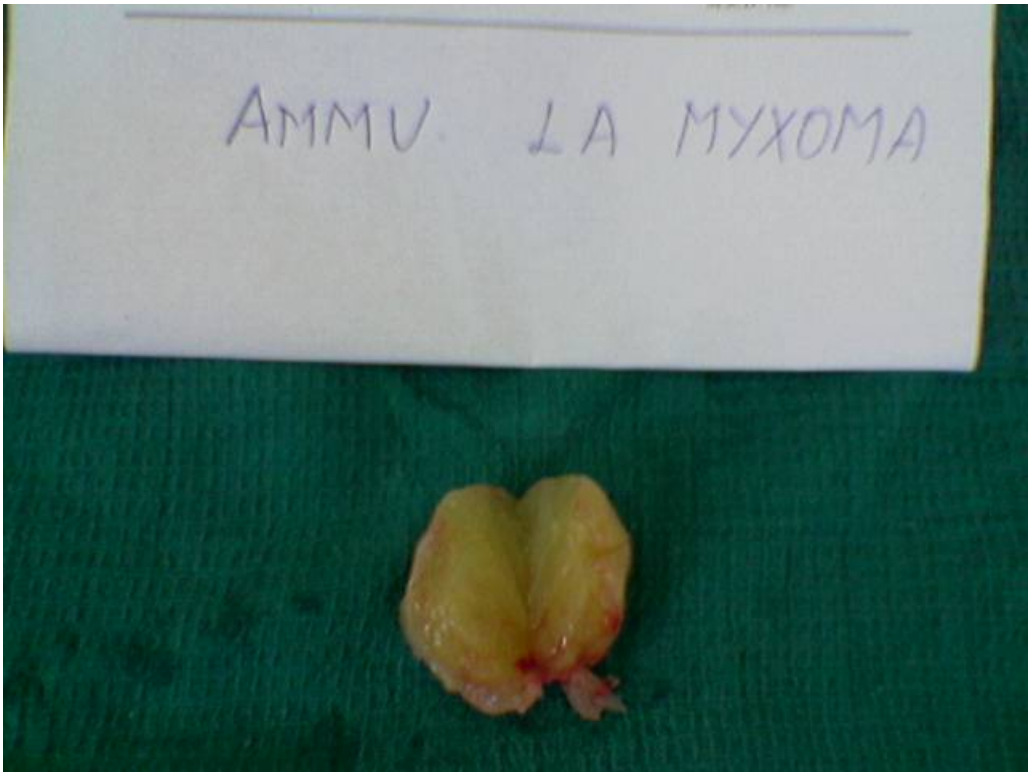
heart to prevent tumour embolization, which can be a serious intraoperative complication of this procedure.

Standard surgical approach for the management of left atrial myxoma is Bi-atrial ,but in our experience in the management of 30 cases, we used Right atrial approach alone for the management of 25 cases of cardiac myxomas. Of the 21 cases of isolated left atrial myxoma without involving mitral valve, 20 cases were approached through right atrium and one case was approached through left atrium. All the four cases of Right atrial myxoma were operated through right atrium. Of the two right ventricular myxomas, one was removed through right atrium and the other required a right ventriculotomy as shown in Table-7. Bi-atrial approach was used in 3 cases were tumour was hugely enlarged and adherent to mitral valve. The mean Aortic

cross clamp time and Cardiopulmonary bypass time was shorter in patients operated through right atrium compared to patients who were operated through Bi-atrial and left atrial approach as shown in table 8. All cases included in the study were discharged and no deaths have occurred. From our study right atrial approach as an alternative to Bi-atrial approach in the management of cardiac myxomas arising from left atrium is worth mentioning.

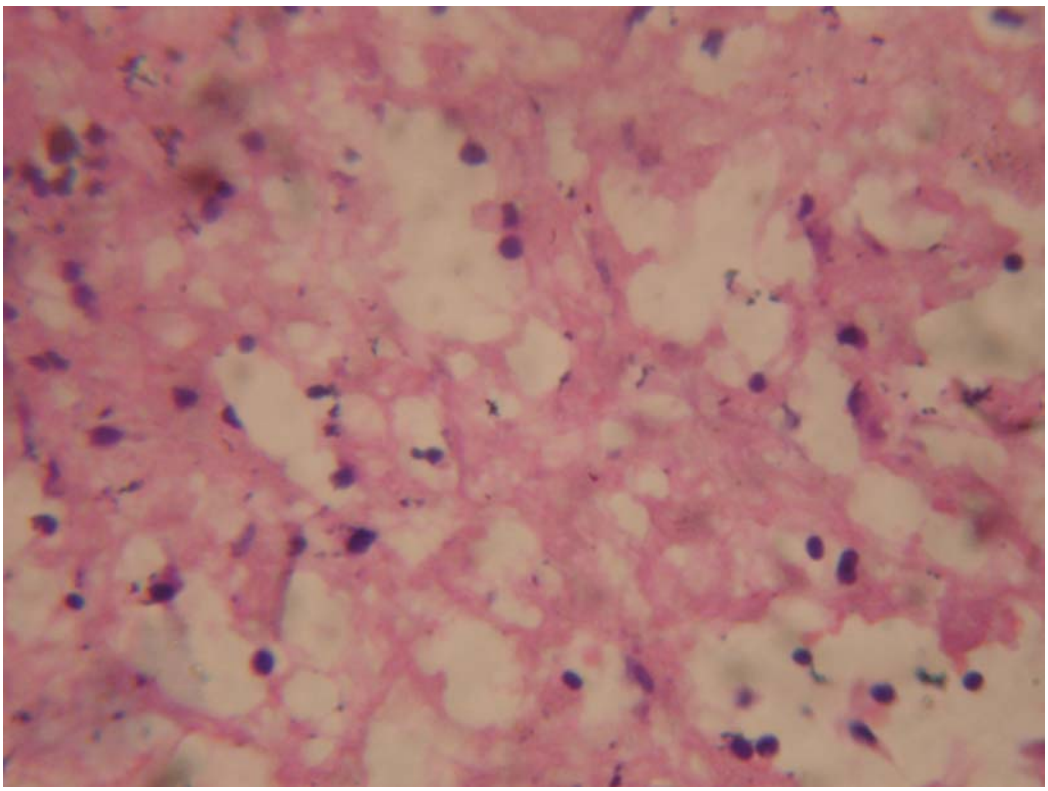


PHOTOGRAPH SHOWING INTRAOPERATIVE TUMOUR REMOVAL THROUGH RIGHT ATRIAL APPROACH.

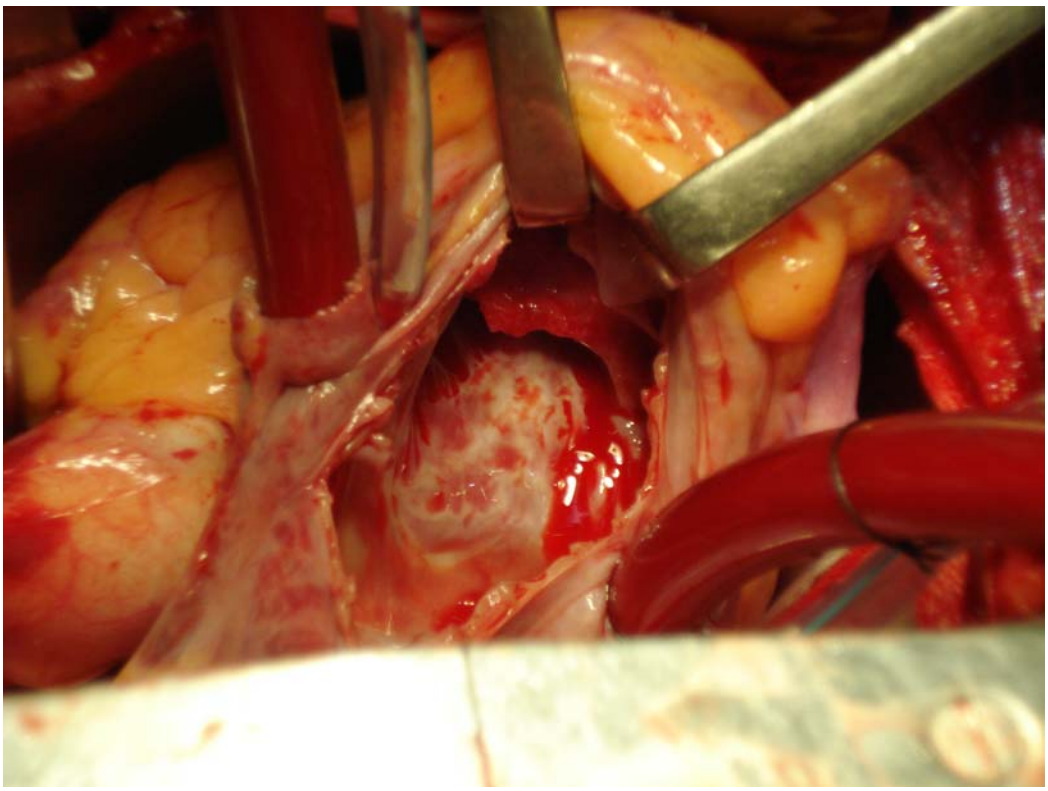


PHOTOGRAPH SHOWING 3x3 CM LEFT ATRIAL

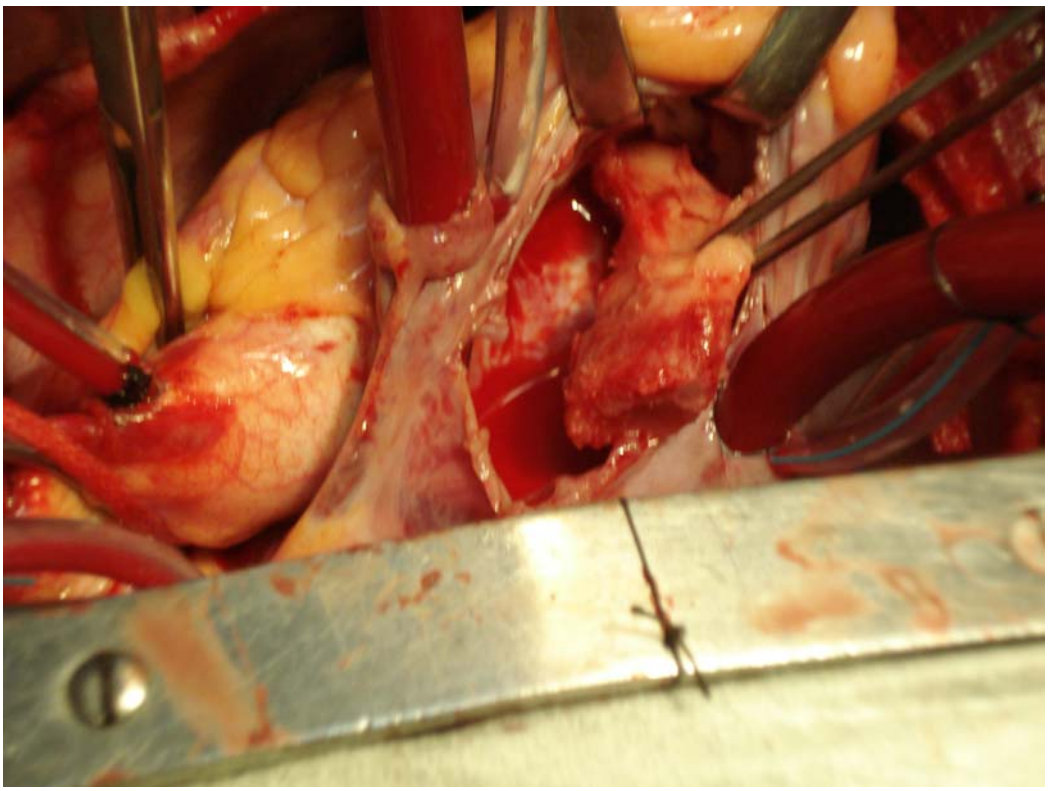
MYXOMA MASS OF A PATIENT



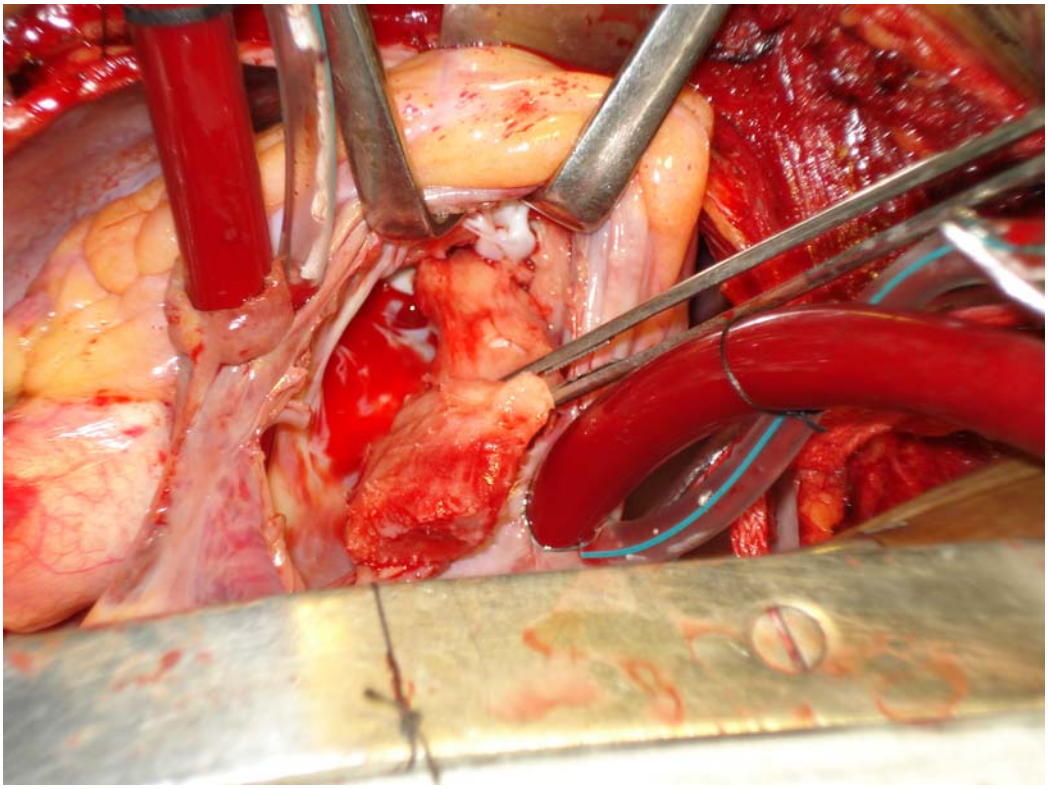
HISTOPATHOLOGICAL PHOTOGRAPH OF CARDIAC MYXOMA



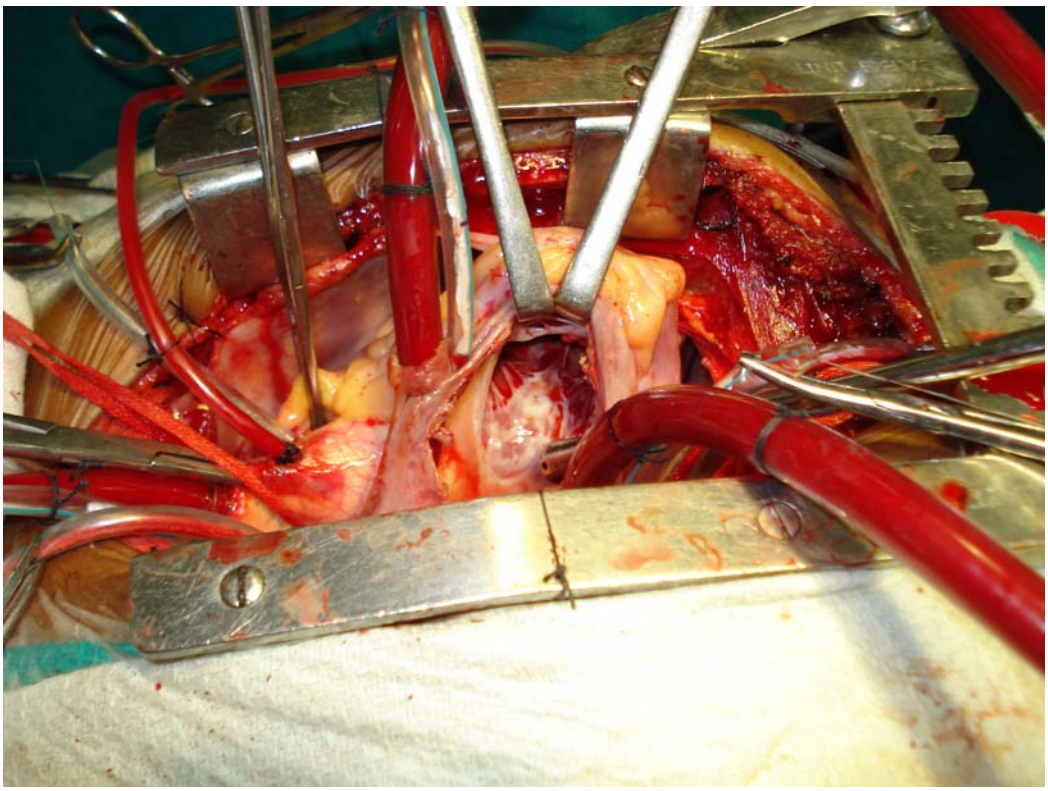
OPERATIVE PHOTOGRAPH SHOWING A RIGHT VENTRICULAR MYXOMA VISUALISED THROUGH THE RIGHT ATRIUM



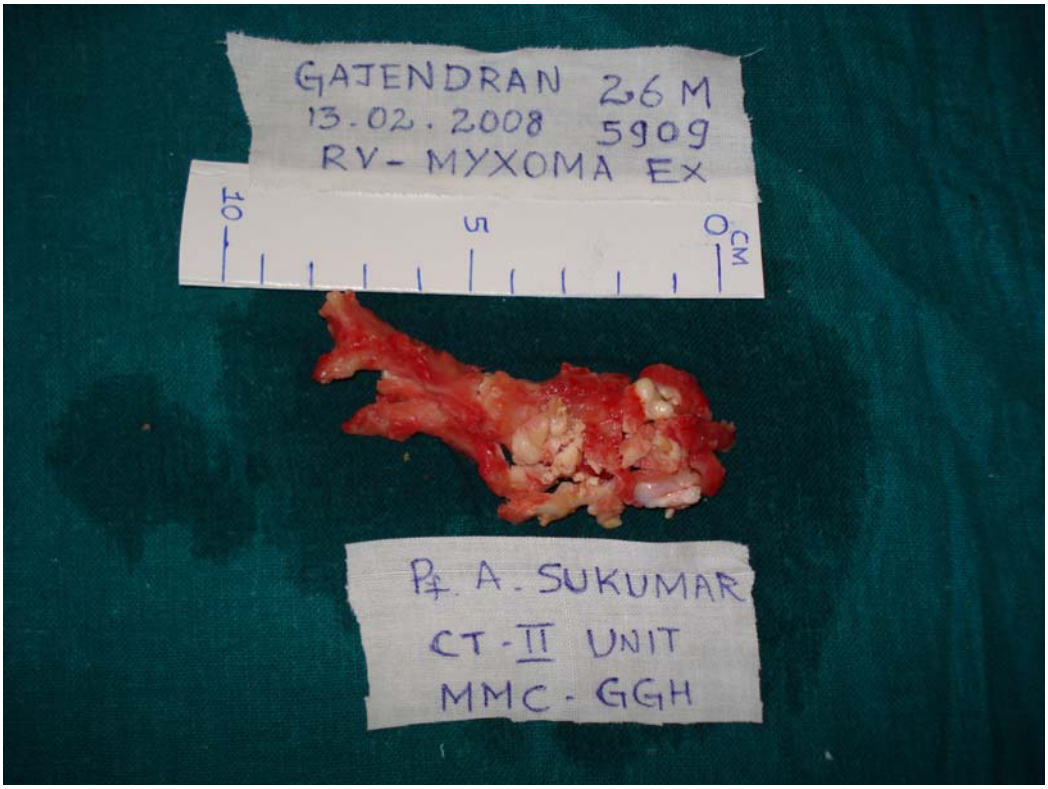
PHOTOGRAPH SHOWING RIGHT VENTRICULAR MYXOMA EXCISED THROUGH RIGHT ATRIUM



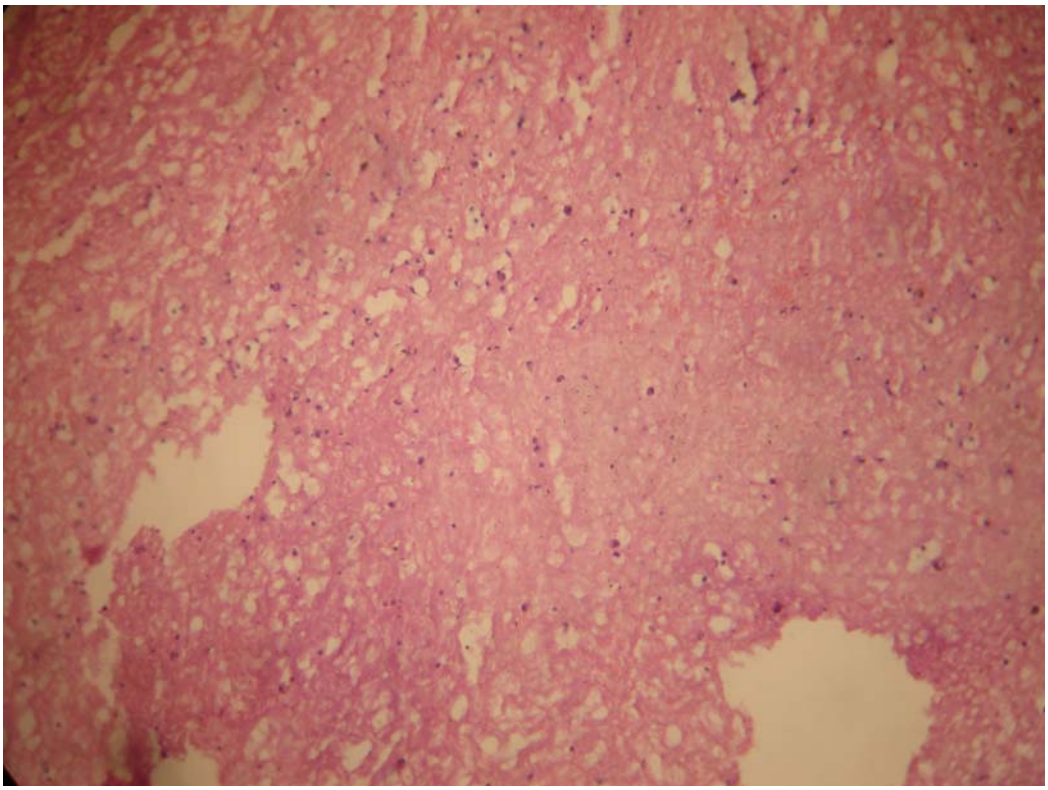
PHOTOGRAPH SHOWING LEFT ATRIAL MYXOMA EXCISED THROUGH RIGHT ATRIUM



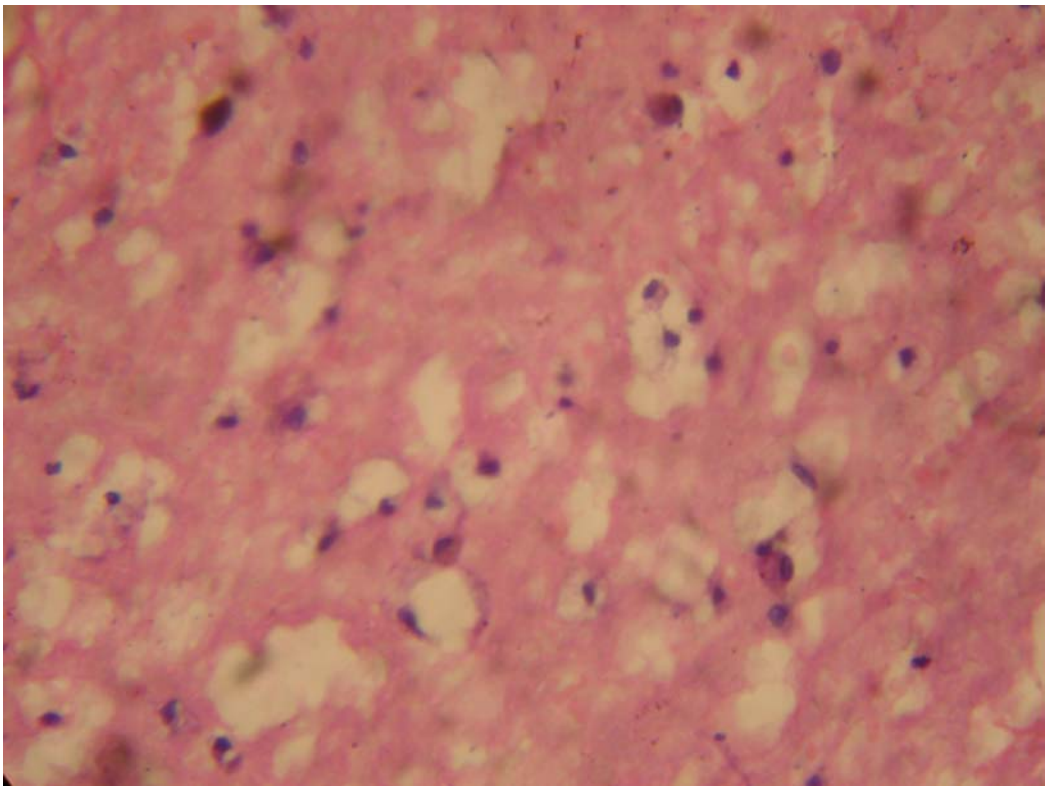
PHOTOGRAPH SHOWING THE INTER-ATRIAL SEPTUM CLOSED WITH PERICARDIAL PATCH



PHOTOGRAPH SHOWING SPECIMEN TUMOUR OF RIGHT VENTRICULAR MYXOMA



HISTOPATHOLOGICAL PHOTOGRAPH OF CARDIAC MYXOMA



HISTOPATHOLOGICAL DEMONSTRATION OF MYXOMA CELLS

Summary and Conclusions

A retrospective analysis of 30 consecutive cases of Intra cardiac myxomas were analysed according to their age, sex , mode of presentation, method of diagnosis, site of the tumour and surgical approach. The surveyed cases showed nearly 73% and 27% of myxomas occurred in females and males respectively. Distribution of myxomas by age indicated nearly 83% of cases occurred between 20-49 years age group. This data also illustrated that 100% males and 77% females harboured the disease between 20-49 years. All the cases included in the study (100%) presented with cardiac symptoms which warranted them for medical attention. However a systematic screening is essentially required to diagnose and treat cases at asymptomatic

level with a view to prevent morbidity. 7% of the cases included in the study presented with neurological symptoms, 27% cases had constitutional symptoms. Trans thoracic echocardiography was diagnostic in 83% of the cases. However, Trans esophageal echocardiography was needed for confirmation in the rest of the cases (17%) .Angiocardiography as a mode of investigation in the diagnosis was not required in our study. In our observation 80% (24 cases) of the tumours occurred in left atrium of which 21 cases (70%) involved the inter atrial septum, 2 cases involved the left atrium and mitral valve and 1 case was diagnosed to have mitral valve myxoma. 13% of the cases were found in the right atrium and 7% cases were observed in the right ventricle. The fossa ovalis was the site of pedicle attachment in 73% of the cases. Right atrial approach was used in 83% of the cases for

tumour excision , Bi-atrial approach was needed in 10% ,left atrial and right ventricular approach was needed in 1 case each. The mean cross clamp time and total cardiopulmonary bypass time was shorter when right atrial approach was employed for tumour excision as compared to classical bi-atrial approach. Hence we conclude that cardiac myxomas are a rare group of tumours and cardiovascular surgeons must be familiar with this condition as only a tip of an iceberg present to us, right atrial approach could be used as an alternative to the classical bi-atrial approach.

Proforma

- ANNEXURE 1
- PATIENT NAME: AGE/SEX:
- MRD NO:
- ADDRESS: OCCUPATION:
- CONSULTANT IN-CHARGE: UNIT:
- DATE OF ADMISSION:
- DATE OF SURGERY:
- DATE OF DISCHARGE:
- CHIEF COMPLAINTS:
- CARDIAC SYMPTOMS:DYSPNEA /PALPITATIONS/ PND/ CHESTPAIN/ PEDAL EDEMA.
- NEUROLOGIC SYMPTOMS:
- CONSTITUTIONAL SYMPTOMS:FEVER/ WEIGHT LOSS/ ARTHRALGIA/
- PAST HISTORY: FAMILY H/O
- GENERAL EXAMINATION:

- PALLOR +/-
- CYANOSIS +/-
- JAUNDICE +/-
- CLUBBING +/-
- PEDAL EDEMA +/-
- PR:
- BP:
- SYSTEMIC EXAMINATION:
- CARDIOVASCULAR SYSTEM:
- RESPIRATORY SYSTEM:
- NEUROLOGICAL EXAMINATION:
- INVESTIGATIONS:
- Hb: PCV: ESR:
- ECG:
- CHEST X-RAY:
- TRANS-THORACIC ECHOCARDIOGRAPHY:
- TRANS-ESOPHAGEAL ECHOCARDIOGRAPHY:

- CORONARY ANGIOGRAPHY:
- DIAGNOSIS:
- SURGICAL PROCEDURE:
- APPROACH: RT ATRIAL/BI-ATRIAL/LEFT ATRIAL
- TUMOUR LOCATION/SIZE:
- SITE OF ATTACHMENT OF PEDICLE:
- CLOSURE OF IAS DEFECT:
- CROSS CLAMP TIME:
- TOTAL CPB TIME:
- OUTCOME:
- BIOPSY REPORT:

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NAME	AGE	SEX	IP NO	DIAGNOSIS	CS	NSS	ESR	CXR	ECHO	CAG	APPROACH	SIZE	SITE OF PEDICLE	IAS CL	ACC	CPB	OUTCOME	BX
KRISHNAN	42	M	611620	LA MYXOMA	+	--	N	N	D	NO	RA	5x5	IAS/FO	PPC	54'	98'	DIS	PROVEN
MEHRUNISHA	35	F	613505	RV MYXOMA	+	--	E	RA/RV EN	D	NO	RV	4x4	TR.SEPTUM	----	66'	122'	DIS	PROVEN
SHANTI	43	F	629835	LA MYXOMA	+	+	N	LAE	D	NO	RA	4x4	IAS/FO	PPC	52'	96'	DIS	PROVEN
MANI	35	M	622978	LA MYXOMA	+	--	N	LAE	D	NO	RA	3x3	IAS	PPC	56'	105'	DIS	PROVEN
PONNAMAL	22	F	634534	LA MYXOMA	+	+	N	N	D	NO	RA	2x2	IAS/FO	DC	34'	52'	DIS	PROVEN
PADMA	42	F	662935	LA MYXOMA	+	+	N	LAE	D	NO	RA	4x4	IAS	PPC	62'	126'	DIS	PROVEN
BRINDA	13	F	656416	LA MYXOMA	+	+	E	LAE	D	NO	RA	7x5	IAS/FO	PPC	58'	128'	DIS	PROVEN

KUPPAMAL	40	F	632098	LA MYXOMA		+	--	N	N	D	NO	BI-ATRIAL	4x4	IAS/FO	PPC	72'	144'	DIS	PROVEN
REVATHY	25	F	701329	LA MYXOMA-MR		+	+	N	CM	D	NO	BI-ATRIAL	5x5	AML/PML/LAROOF	MVR/PPC	112,	156'	DIS	PROVEN
PRABHA	30	F	722189	LA MYXOMA		+	--	N	N	D	NO	RA	3x2	IAS/FO	PPC	48'	92'	DIS	PROVEN
CHELAMAL	40	F	696320	LA MYXOMA		+	--	E	N	D	NO	RA	3x2	IAS/FO	PPC	50'	98'	DIS	PROVEN
DAYALAN	33	M	708292	LA MYXOMA	- MR	+	+	N	LAE	D	NO	BI-ATRIAL	8x5	IAS/ANT.CUSP/COR	MVREP/PPC	118'	160'	DIS	PROVEN
MARIAMA	28	F	712683	RA MYXOMA		+	--	E	N	D	NO	RA	3x3	IAS/FO	PPC	38'	54'	DIS	PROVEN
KRISHNAN	30	M	708828	RA MYXOMA		+	--	E	N	D	NO	RA	3x2	IAS/FO	PPC	36'	54'	DIS	PROVEN

NAVAMANI	36	F	709473	LA MYXOMA	+	--	N	N	D	NO	RA	4x3	IAS/FO	PPC	40'	60'	DIS	PROVEN
KULLAN	36	M	572350	LA MYXOMA	+	--	N	N	D	NO	RA	4x4	IAS/FO	PPC	42'	58'	DIS	PROVEN
BEEMASWAMY	45	M	808679	LA MYXOMA	+	--	N	N	D/TEE	NO	RA	2x2	IAS/FO	DC	32'	50'	DIS	PROVEN
PITCHAMAL	58	F	820422	LA MYXOMA	+	--	N	N	D	N	RA	3x3	IAS/FO	PPC	44'	96'	DIS	PROVEN
MANI	45	F	841824	RA MYXOMA	+	--	N	N	D	NO	RA	6x6	IAS/FO	PPC	55'	100'	DIS	PROVEN
PONNI	42	F	843278	LA MYXOMA	+	--	N	N	D/TEE	NO	RA	4x3	IAS/FO	PPC	56'	106'	DIS	PROVEN
VALLI	27	F	854432	LA MYXOMA	+	--	E	LAE	D/TEE	NO	RA	5x5	IAS/FO	PPC	64'	124'	DIS	PROVEN
MUNIAMAL	53	F	827229	LA MYXOMA	+	--	N	LAE	D	N	RA	5x5	IAS/FO	PPC	58'	114'	DIS	PROVEN

SARAVANAN	20	M	774827	MYXOMA MV	+	+	E	LAE	D	NO	LA	6x6	PML	MVR	108'	148'	DIS	PROVEN
GAJENDRAN	26	M	5909	RV MYXOMA	+	--	N	RAE	D	NO	RA	4x4	RV WALL	----	62'	134'	DIS	PROVEN
SELVI	20	F	27968	LA MYXOMA	+	--	E	LAE	D/TEE	NO	RA	5x5	IAS/FO	PPC	68'	126'	DIS	PROVEN
SUNDARI	43	F	72380	LA MYXOMA	+	--	N	LAE	D	NO	RA	4x4	IAS/FO	PPC	52'	118'	DIS	PROVEN
AMMU	32	F	66553	LA MYXOMA	+	--	N	N	D	NO	RA	3x3	IAS/FO	PPC	40'	64'	DIS	PROVEN
PAPPA	60	F	81543	RA MYXOMA	+	--	N	RAE	D	NO	RA	4x4	IAS/FO	PPC	54'	116'	DIS	PROVEN
DHANALAXMI	40	F	13203	LA MYXOMA	+	+	N	N	D/TEE	NO	RA	4x4	IAS	PPC	58'	94'	DIS	PROVEN
MEENA	18	F	773241	LA MYXOMA	+	--	N	N	D	NO	RA	2x2	IAS/FO	DC	36'	58'	DIS	PROVEN

APPENDIX 2

- KEY TO MASTER CHART
- RA: RIGHT ATRIAL
- LA: LEFT ATRIAL
- RV: RIGHT VENTRICLE
- LAE: LEFT ATRIAL ENLARGEMENT
- RAE: RIGHT ATRIAL ENLARGEMENT
- CM: CARDIOMEGALY
- IAS: INTER ATRIAL SEPTUM
- FO: FOSSA OVALIS
- AML: ANTERIOR MITRAL LEAFLET
- PML: POSTERIOR MITRAL LEAFLET
- PPC: PERICARDIAL PATCH CLOSURE
- DC: DIRECT CLOSURE
- CAG: CORONARY ANGIOGRAM
- MVR: MITRAL VALVE REPLACEMENT
- ACC: AORTIC CROSS CLAMP TIME
- CPB: CARDIO-PULMONARY BYPASS